CLINICAL PROCEEDINGS

of the

CHILDREN'S HOSPITAL

WASHINGTON, D. C.

June 1952

VOLUME VIII NUMBER 6

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CLINICAL PROCEEDINGS

OF THE CHILDRENS HOSPITAL

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OSTEOGENESIS IMPERFECTA

Special Report No. 234

James A. Sowers, M. D.

The objective of this presentation is to review the cases of osteogenesis imperfecta that have been treated as in-patients at The Children's Hospital of Washington, D. C. during the past eleven years, with a description of clinical findings and a discussion of the data obtained.

This disease has been given many names by the many authors that have written of it—i.e., ossium fragilis, osteopathyrosis, Lobstein's disease, "soft bones," and "fragile bones." It is a generalized bone disease occurring primarily in the developing bones of children, but occasionally is not discovered until adult life. It is a congenital-hereditary disease that is characterized, particularly, by the occurrence of numerous fractures which occur spontaneously or with minimal trauma in people who have blue sclerae and occasionally deafness. "About one-third of the cases have a definite hereditary background and occur as a dominant trait—about one-half of the offspring will have the condition, and a generation is not skipped."

Nine patients with the diagnosis of osteogenesis imperfecta have been admitted to Children's Hospital during the past eleven years (see Table I). Although these are relatively few in number, they represent 27 admissions with a total of 371 bed days, and there is good evidence in the records of these patients that several of them have been treated in other hospitals during their life and during this eleven-year period.

In this series of five girls and four boys, one of the girls was colored and the rest of the patients were white. Four of the cases were siblings and two were cousins. A family history of known osteogenesis imperfecta or of multiple fractures was obtained in seven of the patients. The equal sexual distribution is also borne out in these families for in five cases the father was the direct hereditary line and in two the mother was the direct hereditary line. The grandmothers of four and the grandfather of another had apparently had this disease. The family of four siblings, admitted to this hospital, had two other siblings apparently free of the disease, and one with no fractures, but with blue sclerae. In the families affected with osteogenesis imperfecta a history of a number of associated conditions was recorded; i.e., blindness, baldness (since birth), spastic paralysis, deformities causing crippling, and syphilis.

These nine patients have sustained at least 65 fractures during their lives. These fractures have involved mainly the long bones of the upper and lower extremities, although they have occurred in many other bones.

The first fracture occurred in four of these patients at birth, and in one the first fracture did not occur until age seven years. In all but one instance, the admissions to the hospital were primarily for treatment of a fracture; the one exception was admitted for treatment of meningitis.

TABLE 1

CASE	HOSP. NO.	RACE SEX	NO. ADM. TO HOSP.	FIRST FRACT. AGE	DIAGNOSIS MADE AT AGE	BLUE SCLERA	FOLLOW UP
S. P.	41-3588	WF	1	6 yrs.	?	Present	Walks with limp
R. W. P.	41-7950	WM	5	14 mos.	10 yrs.	Present	Ankylosis, elbow
H. P.	41-7951	WF	2	7 yrs.	7 yrs.	Present	No deformities
G. P.	42-1238	WF	1	Birth	?	Present	Died meningitis
S. L. F.	45-7727	CF	10	1 mo.	7 yrs.	Present	Limps, muscle atrophy, can't walk upstairs flexion contr. hips
P. W. B.	46-9356	WM	1	Birth	3 wks.	Present	Died of pneumonia with multiple congenital deformities
H. F. R.	48-13697	WM	3	Birth	1 mo.	Present	Bowing of legs & shortening muscle atrophy
E. M. S.	49-7796	WF	6	Birth	18 mos.	Present	No deformities
D. H.	49-11579	WM	1	1½ yrs.	1½ yrs.	Present	No deformities

Chart 1 clarifies the distribution of the fractures that have occurred. The types of fractures involved are varied, but it is notable that many of them were spiral fractures and none of them were compound. Many deformities have resulted from the fractures, especially in those with multiple fractures. They include such things as ankylosing of a joint, bowing of the bones, contractures of the hips, atrophy of the muscles of the extremities,

and inability to walk well or at all. Most of the fractures that occur were the direct results of falls of minor nature. A number of them were from such minimal trauma as twisting or being lifted.

Because of the large number of fractures, a total of over 200 X-rays have been made. The most common findings, other than that of multiple fractures, that indicated a diagnosis of osteogenesis imperfecta to the radiologists were; osteoporosis with narrow diameter of the bones, flaring of the ends of the long bones, and thinning of the cortex; also rarified areas representing pseudocystic areas, which occurred in some of the long bones

CHART 1

Distribution of Fractures

CASE	LOCATION OF FRACTURE																		NO. FRACTURES
	Upper Extremity							Lower Extremity							Other				
	Arm	Forearm						Femur		Tibia		Fibula							
	Not spec.	Radius		Ulna		Elbow	Not spec.							Not spec.	Ribs	Vertebra	Scapula	Clavicle	
		L	R	L	R			L	R	L	R	L	R						
1			2		2		1			1	1	1	1	—					9
2	4	2		3		1								3					13
3		3		3															6
4																			
5								2	3					5			1		11
6		1		1												2	1		5
7									8	1					4				13
8								2	1	3								1	7
9									1										1
Total	4	6	2	7	2	1	1	4	13	5	1	1	1	8	4	2	2	1	65
	23							33							9				

that were fractured. Such conditions as osteomalacia, periostitis and periosteal proliferation, hyperostoses, and fragmentation of the intermedullary trabeculations were also notable findings. Two of the cases presented definitive findings on X-rays of the skull—one showed a mosaic pattern of the bone and the other had platybasia. Another patient had micrognathia. Though X-rays provided a valuable adjunct, in none of the cases did it make the diagnosis without the aid of clinical evaluation.

The most common presenting symptom in these cases was fracture, but only two of them were diagnosed at the time of their first fracture and several of them were not diagnosed as having osteogenesis imperfecta

until they had had numerous fractures. The most common first clue to the diagnosis was the presence of blue sclerae. All nine of these patients were observed to have blue sclerae, and dental changes were a prominent feature of several of them—such as greenish deformed alveolar ridges at birth, and later bluish teeth with soft enamel, which in some instances were worn down to the gums. Partial deafness was observed in one, and deafness of bone and nerve conduction in another case; but this latter may have been the sequela of pneumococcic meningitis.

Many times osteogenesis imperfecta was confused with other possible diagnoses; such as hyperparathyroidism, congenital or renal rickets, scurvy, bone tumors of the giant-cell cyst type, metastases of cancer, osteitis fibrosa cystica congenita, neurofibromatosis of the bone, and osteomalacia. Conditions other than those related to osteogenesis imperfecta were sometimes present. Some of these have been mentioned. One child had myopia, another pneumococcic meningitis, another severe eczema, another pigeon-breast, and another congenital rickets. Three of them had vaginitis—one of these was due to gonorrheal infection. Thus often these cases presented double problems of care.

These nine cases have required numerous procedures of treatment. Bryant's traction with a Bradford frame on an orthopedic bed was used in practically all cases of fractured femurs under seven years of age and Russell's traction has been applied in the care of those over this age. Closed reductions of the fractures with immobilization was necessary in almost all of the cases though in some the displacement of the fragments required only minimal manipulation. Consequently, well over sixty plaster casts and splints were applied. In the majority of fractures, casts had to be reapplied several times. Large casts, such as hip spicas, were necessary in practically all of the femur fractures. Two open reductions were performed with wiring of the non-united fragments, and subsequently these wires were removed because of infection of the bones. Numerous braces, special shoes, and other orthopedic appliances had to be used in the follow up care. A great deal of nursing care was necessarily given and the children received antibiotics, physical therapy, analgetic drugs, social and psychiatric guidance, and various supportive treatments—all of which signify a large amount of time and money involved in the care of these patients. In addition many were unable to attend school for long periods and failed to receive instruction at home.

Very little laboratory work other than routine was done. However, three cases had calcium and phosphorus level determinations in the blood and the results were normal. One case had an alkaline phosphatase of ten King-Armstrong units. The urine total calcium in one twenty-four hour specimen was 16.6 mgs. per 100 ml.

A bone biopsy was performed on a benign cyst and one autopsy was done. The bone changes that were found in autopsy revealed the cartilagenous bone to have irregular islands of calcification, most marked in the flat bones. The bones of the extremities were soft, could be crushed between the thumb and fingers and were described as feeling like corrugated paper. It was noted that the blood flowed freely from the bones as they were cut with a knife and, also, that the bones of the extremities all showed irregular bowing.

GENERAL DISCUSSION

From the data obtained on these cases it becomes apparent that the amount of man-hours and materials required in the care of these patients who are afflicted with osteogenesis imperfecta, is tremendous. It is indeed fortunate that the cases are uncommon. Though there are only nine cases listed here at least three others have been treated as out-patients in the Orthopedic Clinic. Each fracture that occurs must be treated carefully to prevent deformity, and despite careful treatment deformities still necessarily result. Some are severe enough to make these children crippled for life. Fortunately the number of fractures seems to decrease as the child grows older.

The exact etiology of this disease is not clear and attempts to ascertain if it is of endocrine origin have apparently failed. Although it is difficult to diagnose this condition without the appearance of multiple fractures, it is well to be on the alert in all cases of fracture for a family history of the disease as well as to observe the hue of the sclerae and the conditions of the dentition. It is desirable that osteogenesis imperfecta be diagnosed early so that, with careful discipline at home, some fractures can be prevented—for apparently fractures recur very frequently in those bones that have previously been fractured.

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POTASSIUM DEFICIENCY

Case Report No. 235

Joseph M. LoPresti, M. D.
Francis J. Mastrota, M. D.
David L. Simon, M. D.

INTRODUCTION

In the past few years of rapid medical progress, it has become more apparent to the practitioner that almost all disease processes are attended by fluid and electrolyte imbalances. This altered physiology in disease may be a major factor in the production of the clinical symptomatology. A recognition of this fact together with corrective therapeutic measures are essentials to rapid recovery and a favorable outcome. However, both the biochemist and clinical laboratory have labored under physio-chemical limitations. Direct measurements are possible only on the easily accessible extracellular compartment of the human body. While sodium, bicarbonate, and chlorides are present in large amounts in this compartment, electrolytes such as potassium, calcium, magnesium, and phosphates occur in relatively minute fractions. A major altered physio-chemical state may be reflected by only minor changes in the extracellular content of these "trace" substances. Until recently, the importance of these minor changes in the extracellular fluid has been overlooked. The errors inherent in the determination and interpretation of results obtained by chemical analysis of the extracellular compartment obviously are responsible for this belated recognition.

A wealth of medical literature has accumulated concerning the role that potassium plays in pathological states. It is the purpose of this discussion to summarize the more important aspects of potassium metabolism, with particular emphasis on the etiology, symptomatology, diagnosis, and treatment of potassium deficiency.

CASE REPORT

The following case is presented as being illustrative of one of the more common pediatric conditions which may produce potassium depletion.

A. B., a six-week old colored male, was admitted to the diarrhea service with a history of 12 to 15 watery green stools for 24 hours. Physical examination revealed a poorly nourished, markedly dehydrated, colored male. The anterior fontanel was depressed; skin turgor was poor and the infant appeared weak and listless. The temperature was normal. Red blood cells numbered 3,000,000; hemoglobin, 10 grams; CO_2, 32 volumes per cent. The infant was treated with intravenous 1/6 molar sodium lactate, 5 per cent glucose in Hartman's solution, blood transfusion, and 30 milliliters of Darrow's solution by mouth every 4 hours. As he improved he was given a formula of powdered protein milk.

Electrocardiogram taken on admission (Fig. 1) showed the heart rate to be 140 per minute. The cycle length was 0.44 seconds. The Q-T internal was greater than 0.36 seconds. The T wave showed a slow upward swing in the S-T segment and low voltage. The end of the T wave ran into the following P wave. The electrocardiogram taken 5 days later showed a rate of 140 per minute. Cycle length was 0.44 seconds. Q-T internal was 0.28 seconds with a normal configuration of the T wave. The patient was clinically improved at this time and was discharged shortly thereafter in good condition.

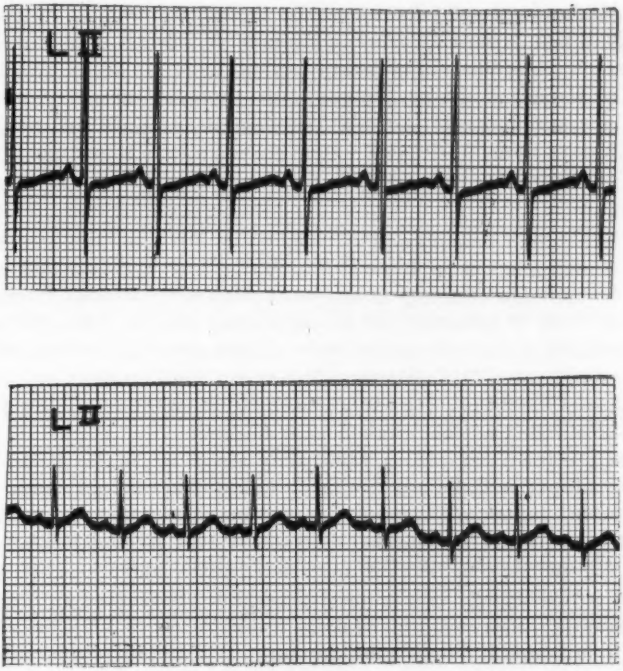

FIG. 1. Electrocardiographic tracings on the day of admission and five days later. (Upper) ECG on admission, lead II. (Lower) ECG 5 days later, lead II.

PHYSIOLOGY

The human body contains from 175 to 250 grams of potassium; an amount sufficient to kill 30 people and represents three times the sodium content. However, the normal plasma level of potassium is only 5 milliequivalents per liter (20 milligrams per 100 milliliters), while the normal value for plasma sodium is 142 milliequivalents per liter (617 milligrams per 100 milliliters). The remainder of the body potassium (97 per cent) is located within the intracellular compartment, primarily in the tissue cells and

erythrocytes. Two-thirds of the intracellular potassium is bound to protein and one-third of it diffuses. Potassium enters the cells during protein and carbohydrate anabolism; it leaves the cells during tissue breakdown and in the presence of alkalosis. In the normal individual 3 to 4 grams of potassium are ingested and excreted each day.

The intracellular concentration of potassium can fluctuate without an appreciable effect on the human organism. However, even small changes in the plasma level will effect markedly the physio-chemical status of an individual and produce clinical symptoms. A normal concentration of potassium in the plasma is necessary for:

1. The activation of acetylcholine at the parasympathetic synapses.
2. The transmission of electrical impulses along nerves and across the myoneural junctions.
3. The myocardium is extremely sensitive to variations in plasma potassium levels.

Pathologic Physiology

The human organism may be effected profoundly by either excessive or deficient levels of potassium in the circulating plasma. The mechanisms by which greater than normal amounts of potassium will be found include: (1) the rapid parenteral administration of potassium-containing solution, e.g., Darrow's solution; (2) and pathological states in which renal function is impaired (particularly glomerular filtration).

There are many conditions which may result in potassium depletion: (1) an inadequate intake, e.g., bulbar poliomyelitis; (2) an increase in the excretion of potassium related to an increase in nitrogen excretion, e.g., infection, neoplasm, uncontrolled diabetes, starvation, and post-operative states; (3) chronic renal disease in which excessive amounts of potassium are lost in the urine; (4) prolonged periods of vomiting, e.g., intestinal-obstruction; (5) measures which are instituted to relieve abdominal distension or vomiting, e.g., Wagensteen drainage; (6) diarrhea in which excessive amounts of base are lost in the feces; (7) conditions in which a shift of potassium from the plasma into the cells occurs, particularly during nitrogen and carbohydrate anabolism, e.g., the administration of insulin, testosterone, or cortisone, and periodic paralysis.

SIGNS AND SYMPTOMS

The clinical picture presented by a patient suffering from potassium deficiency is a well defined and easily recognizable entity. There will be present clinical conditions capable of producing the deficiency, e.g., diarrhea, intestinal obstruction, renal disease, etc. If a mild depletion is present, the patient will be dehydrated and listless. Chronic ileus commonly will

be seen. Edema and oliguria will be present. A prominent feature will be loss of strength and energy, e.g., the patient will be unable to feed himself. If the deficiency is more marked or uncorrected, weakness of the extremities will develop and a flaccid paralysis of the Landry's ascending type may occur. Tachycardia will be present. There will be weakness of the respiratory muscles and the respirations will be rapid and shallow. Air hunger, in which the patient utilizes his accessory muscles of respiration, may be a prominent feature. There will be dilation of the alae nasi and "fish-mouth" breathing. The chest wall and diaphragmatic excursions will be diminished. Ultimately, complete respiratory paralysis may occur.

DIAGNOSIS

The diagnosis of potassium deficiency may be established readily by characteristic electro-cardiographic changes and laboratory determinations of the plasma-potassium level.

In the presence of a low potassium content, the electro-cardiogram will show a delayed A-V conduction time; in the presence of moderate decreases, the T waves will be lower and broader than normal and the Q-T interval will be prolonged; with further decreases, the T waves will be low and flat and may become inverted, and there will be sagging of the S-T segments.

The most rapid and accurate determinations of potassium levels in the plasma will be furnished by the flame photometer.

TREATMENT

The parenteral use of a potassium-containing solution to correct a deficiency of this element is not without danger. *The solution should never be used in the presence of oliguria, hemoconcentration, or renal disease.* The dose of potassium is 0.13 grams per kilogram of body weight. When administered parenterally, it should be injected slowly over a four to eight hour period. It must be kept in mind that the risk of intoxication is ever present. By far, the best route is the oral one. Three to five grams of potassium chloride may be added to a liter of milk or fruit juice. Orange juice has a high content of potassium and may be utilized in moderate amounts.

SUMMARY

The physiology, pathology, clinical symptomatology, diagnosis and treatment of potassium deficiency has been discussed briefly. A case report illustrating one of the conditions responsible for potassium depletion has been presented.

A REPORT OF A CASE OF ATLANTO-AXIAL DISLOCATION IN A MONGOL

Case Report No. 236

John M. Kennelly, Jr., M. D.

M. S., a four year old white female mongoloid, was admitted to the hospital on February 6, 1952 with the history of having fallen on her buttocks and striking the back of her head on the ground the day prior to admission. Since then she had been fretful and had slept fitfully. There was limitation of motion of all extremities without actual paralysis. She developed gasping respirations, but no dysphagia was noted.

The past history revealed the usual retarded development of the mongoloid child. She walked at 2½ years of age and her speech consisted of words with a few phrases. She had been taking glutamic acid, 1 tablespoon daily and thyroid 0.1 grains twice daily. Two years previously she had fallen and suffered a neck injury with similar symptoms—but without the gasping respirations. At that time, x-rays were taken and there was a questionable diagnosis of cervical vertebral fracture, the exact level of the fracture not being known. A neck brace was employed and improvement was gradual. Ten months prior to the present admission she again fell and developed transient limitation of motion of her extremities.

Family history revealed the mother, father and an eleven-year old female sibling living and well.

Physical examination revealed a well developed and well nourished mongoloid child with gasping respirations. Temperature, pulse and respirations were within normal limits. The blood pressure was 90/55. There was localized tenderness on palpation, and pain on flexion of the neck but no nuchal rigidity. Examination of the heart revealed a marked apical systolic murmur. The extremities exhibited marked hypermotility of the joints and weakness which was more marked in the left arm. The neurological examination revealed right ankle clonus, hyperactivity of the deep tendon reflexes in the right lower extremity and absent reflexes in the left upper extremity. Babinski reflexes were negative and sensation was intact.

The clinical impression at this time was cervical vertebral dislocation, probably at the atlanto-axial joint, with spinal cord compression.

CLINICAL COURSE

During the night following admission, the patient developed two attacks of cyanosis and required oxygen therapy. It became evident that there was no intercostal respiration.

LABORATORY

Spinal tap on the day following admission was normal. Blood count and urinalysis were also normal. X-ray examination of the cervical vertebrae revealed an atlanto-axial dislocation with an anterior displacement of the atlas on the axis. The distance between the posterior atlas and the odontoid process of the axis is usually between 18 and 19 min. In this case the distance was 5 min. indicating rather severe cord compression.

A neurosurgical consultation was obtained on February 10, 1952 and the following findings were noted: generalized flaccidity of the muscles more marked on the left, absent reflexes in the upper extremities and hyperactive reflexes in the lower, sensation present thruout and bowel and bladder function normal. Conservative treatment for the next few days was advised, to be followed by returning to the brace wearing or a surgical procedure involving removal of the arch of the atlas.

For the next few days the child was in and out of the respirator and it soon became apparent that her improvement was limited. It became impossible for her to breathe without the aid of the respirator beyond a few minutes time.

SURGERY

Surgery was decided upon and on February 15, 1952 was carried out. A cervical laminectomy with removal of the posterior arch of the atlas, opening of the dura and an occipital craniectomy to enlarge the foramon magnum were performed. The spinal cord was compressed between the adontoid process of the axis and the posterior arch of the atlas prior to the decompression.

Post operatively the child had a very stormy course being able to remain outside of the respirator only for short intervals. She developed considerable tenacious material in the respiratory tree which required frequent aspiration. On 2–18–52 a direct laryngoscopy and bronchoscopy was performed and no obstruction was found. The child went into coma late on 2–18–52, failed to rouse and was pronounced dead on 2–19–52.

AUTOPSY SUMMARY

Ruth H. F. Lau, M.D.:

1. Compression atrophy of cervical cord.
2. Syringo-myelia and demyelinization of the cervical cord seondary to compression.
3. Pulmonary emphysema.
4. Interstitial edema of the myocardium.
5. Acute passive congestion of the viscera.

Gross examination of the cervical cord revealed an area of contusion on the ventral surface situated 1.5 centimeters below the inferior pole of the olives. This contusion measured 0.7 centimeters in diameter and appeared to be slightly depressed and bluish-purple. An angular depressed area was found on the dorsal surface of the cervical cord at the level where laminectomy was performed. The distal segments of the cord was normal in appearance and cross section.

Microscopic examination of the cord at the level of contusion revealed an extensive distortion of the usual architecture. The tissues are canalized in many places, the largest of which is found in the center appearing to be an area of syringo-myelia, being lined by a single layer of fibrocytes. There is marked proliferation of the capillaries carrying intact erythrocytes. The neurons have lost their structural details appearing as hyalinized dots. The nerve fibers present a reticular arrangement suggestive of demyelinization. The pia is thickened, consisting of increased fibrous tissue.

DISCUSSION

John M. Kennelly, Jr., M.D.:

Atlanto-axial dislocations may result from trauma, infection, paralysis or congenital defect.[1] Usually those cases resulting from trauma are associated with a fracture of the odontoid process of the axis.[2] Watson Jones[3] considered that the dislocation alone was more serious than fracture dislocation "because if the odontoid process is intact, the spinal cord is in danger of being crushed against it. The distance between the odontoid process and the posterior arch of the atlas is usually between 18–19 millimeters, and in the case presented, this distance was reduced to 5 millimeters. This posterior pressure of the odontoid process produced a compression of the cervical cord leading to bilateral lower motor neuron paralysis involving respiratory movement.

Although such a condition is usually associated with trauma the nontraumatic atlanto-axial dislocation must be kept in mind. In 1830, Bell[4] described the occurrence of dislocation of the atlas because of destruction of the transverse ligament that holds the odontoid process of the axis in its normal anterior position. This case was associated with a neck infection in the patient. In fact, the otolaryngologic literature classifies such a spontaneous dislocation following cervical infection such as sinusitis, pharyngitis, tonsillitis, mastoiditis or adenitis as Grisel's disease. In these cases, the dislocation is secondary to a destruction of the ligamentous insertion due to calcium decalcification on the basis of calcium absorption from the hyperemia induced by the infection.

In the case presented, the muscular hypotonia and the laxness of the

periarticular ligaments found in the mongol were felt to be the predisposing factors with the trauma of the fall being secondary.

For years this condiion was treated by non-operative means and this is still the treatment of choice in many instances.[5] However, if there is obvious cord compression causing neurological disturbances and endanger ing life, the operative approach is indicated. The first operative attempt to reduce an atlanto-axial dislocation that is recorded was by Lewis Stephen Pilcher[6] in 1900. He was unsuccessful in reducing the dislocation. Mixter and Osgood, in 1912[7], reported two cases in which an open reduction was successfully performed. Foerster[8] in 1922 reported a case of progressive atlanto-axial dislocation in which he removed the arch of the atlas, lamina of the axis and posterior lip of the foramen magnum.

This procedure has been carried out with success since that time. Usually a fusion of the cervical vertebrae to the occiput is performed, but this can be delayed until a later date when the patient's condition will permit such a procedure.

In the case presented, the cord pressure had caused irreversible changes so extensive that recovery was impossible.

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CLINICO-PATHOLOGICAL CONFERENCE

Directed by: E. Clarence Rice, M. D.
Assisted by: Ruth H. F. Lau, M. D.
By Invitation: Morris Michael, M. D.

Ruth H. F. Lau, M. D.

This nine-month old white female was admitted to Children's Hospital on February 20 and died on February 22. The patient was the product of a full-term gestation. Her birth weight was 9 pounds. Respirations were stated to have started spontaneously and the neonatal history was uneventful. The patient was fed on an evaporated milk formula with supplementary vitamins and baby food. She had been in excellent health with no history of any childhood illnesses until four days prior to admission at which time she began to gag and vomit three to four times a day. Following this, slight respiratory distress occurred. This condition became progressively worse until the day before entry to the hospital when the baby developed fever and moderate respiratory difficulty was noted. She was then seen by a local physician who prescribed some "white medicine" and penicillin. The following morning, the baby ran a high fever, was markedly dyspneic and had an excess of frothy sputum.

On admission, the child was pallid and slightly cyanotic and appeared to be in marked respiratory distress. The temperature was 106.0 F. The infant was well developed and well nourished. The fontanel did not bulge nor pulsate. Some viscid white mucus was noted in the mouth and throat. Laryngoscopy failed to reveal any foreign body. The heart rate was 160 per minute and the lungs were filled with fine rales and rhonchi. The respiratory rate was 100 per minute and intercostal retraction was severe. The liver was palpable 4 centimeters below the right costal margin. Roentgenograms were not taken. The provisional diagnosis was acute bronchopneumonia and bronchiolitis.

The family history revealed that the mother was ill in another hospital because of a spontaneous abortion at the time of this child's admission. The father and one other sibling were reported to be in good heath. There was no history of tuberculosis, diabetes or cancer.

A blood count taken on admission revealed 8 grams of hemoglobin; 3,500,000 erythrocytes; 11,800 leukocytes with 65 per cent segmented cells, 5 per cent young cells, 2 per cent myelocytes, 28 per cent lymphocytes, 5 per cent monocytes, and 14 per cent basophiles. The neutrophiles showed toxic granulation. Blood culture showed *B. subtilis*, and throat culture produced normal flora.

Immediately following admission, the patient was placed in an oxygen tent and measures were taken to lower the temperature. Digitoxin 0.1 milligram was given intravenously on the day of admission and was repeated by the intramuscular route the following day. Penicillin (1,000,000 units initially and 500,000 units every 4 hours intramuscularly) and chloromycetin 1 gram twice daily were administered. These procedures caused apparent improvement during the next 24 hours with slight lowering of the respiratory rate to 80 per minute and slight reduction in the number of rales in the chest, however, during the night, the patient became restless, markedly dyspneic, and progressively cyanotic until she expired 45 hours after admission.

DISCUSSION

Morris Michael, M. D.

We have here a previously healthy nine-month old child, whose entire picture is that of respiratory difficulty. Even the onset with gagging and vomiting is all too common in a child struggling with postnasal discharges.

The acute onset and the age of nine months seems to rule out the more esoteric congenital conditions causing respiratory difficulty, such as cystic fibrosis of the pancreas, tracheo-esophageal fistula, and obstructive bands or vessels about the trachea.

The picture is indeed that of an overwhelming, rapidly-fulminating respiratory infection with the excess mucus and plugging of the bronchioles gradually leading to a shock-like state from anoxia. Laryngoscopy failed to reveal a foreign body although presumably this could have been deeper in the respiratory tree. Such a foreign body in the lower trachea, with a ball-valve action could have started a chain of events which, with secondary infection, could not be differentiated from this terminal picture. Vomiting and aspiration of milk could also have lead to aspiration pneumonia and atelectasis.

The blood picture showed a secondary anemia, the leukocyte count indicating an infection. Blood culture revealed what was probably a contaminant, which, incidentally, may have overgrown a pathogenic organism. Throat culture was of no avail, but many of the most virulent organisms such as Hemophilus influenzae are sometimes difficult to grow. Although a primary cardiac picture may certainly cause pulmonary congestion of this degree and enlargement of the liver, it would seem strange that no previous suspicion of cardiac disease had been entertained in a child who obviously had had good care.

It is easier to assume that the child had a secondary myocarditis, due to overwhelming pulmonary infection. The enlargement of the liver could have been merely a liver pushed down by a child exhibiting marked retraction. Almost any severe infection may cause myocarditis. Certainly in diseases such as diphtheria and scarlet fever, we may find electrocardiographic evidence of myocardial disease in 50 per cent of cases and can assume that the other 50 per cent would have clinical evidence of the same were our diagnostic method delicate enough. The use of digitoxin was certainly indicated here.

One wonders what caused the coincident abortion in the mother. I thought of brucellosis, which in animals certainly causes abortion, of toxoplasmosis, and of peptic abortion. Although brucellosis may cause pulmonary congestion, the course is usually mild in children and more chronic.

An organism capable of causing sepsis such as the streptococcus, in the dust of the home, may well have initiated the disease. I consider as unlikely such rare diseases as typhoid pneumonia, tuleremia pneumonia, psittacosis, and plague, but I feel drawn to a consideration of three bacterial organisms and one virus, viz. Hemophilus influenzae, Friedlander's bacillus, and the streptococcus and the virus causing primary atypical pneumonia.

Poisoning must also be mentioned as being capable of causing the development of pulmonary edema and circulatory collapse. Aspirin intoxication may lead to hyperpnea and hyperpyrexia, cyanosis and collapse.

Of all these etiologies certainly bronchiolitis caused by Hemophilus influenzae is most common and most closely fits this clinical picture with its generalized obstructive emphysema, patchy areas of atelectasis, necrosis of the bronchioles and bronchi, causing such severe respiratory distress, that the child uses every accessory muscle of respiration, gradually becoming exhausted and finally asphyxiated.

PATHOLOGIC DISCUSSION

Ruth Lau, M.D.

This case has been presented in its clinical entirety. The findings at autopsy were most interesting to those who followed this patient's illness.

The main pathological findings were in the mediastinum. An open safety pin was found protruding from the posterior wall of the esophagus through an 0.2 centimeter perforation. A retroesophageal abscess was present, measuring 3.0 by 2.0 centimeters, being bounded anteriorly by the esophagus, posteriorly by the vertebral column, and laterally by the great vessels and neck muscles. Pinkish-yellow thick material filled the cavity. The esophageal lumen was empty except for the protruding portion of the safety pin. The mucosa surrounding the perforation within a radius of 2.0 centimeters was slightly edematous. The lungs showed congestion without evidence of consolidation and the apical pleura of the right lung was adherent to the parietal pleura.

Microscopically, the wall of the esophagus, at the area of perforation, was edematous, acutely inflamed, and necrotic. In this area the mucosa had desquamated. In other sections, from the sites of chemical reaction, fibrosis was prominent. The pleura showed acute inflammatory changes and sections of the lungs disclosed congestion of the blood vessels with minimal evidence of pneumonitis. Other findings at postmortem examination were not remarkable except for acute inflammatory changes in the thyroid and central congestion of the liver with fatty metamorphosis.

In summary the pathological diagnosis were:

1. Retroesophageal abscess secondary to esophageal perforation by a foreign body (safety pin) with esophageal ulceration.
2. Thyroiditis secondary to No. 1.
3. Acute pleuritis.
4. Acute congestion and fatty metamorphosis of the liver.

E. Clarence Rice, M. D.

This infant's illness should be of considerable practical importance to us. The history of a foreign body lodging in the esophagus is usually a rather clear cut one, and with our hindsight, it now seems quite typical. If a history is carefully taken, one usually learns of sudden difficulty in swallowing or breathing. Possibly greater attention would have been paid to the onset of this baby's illness had the mother not been absent from the home, the patient having been left in the care of a relative. While a foreign body can sometimes be visualized by laryngoscopy, so little of the pin projected into the lumen of the esophagus that visualization was unlikely. It is possible that the physician in attendance may have considered the patient to have been too ill to be x-rayed. Had roentgenographic examination been done, the diagnosis would have been made. Esophagoscopy in this patient, in all likelihood, might have demonstrated the foreign body. This is not always the case, however, as we have performed autopsies on some children in whom the esophagoscope apparently caused an esophageal perforation by pushing an unrecognized foreign body which was lodged in the wall through it. Prompt cognizance of the nature of the accident and x-ray examination of the foreign body with vigorous antibiotic therapy could prevent the death of such patients. It is unlikely that this patient would have survived the infection which was present at the time of her admission had the pin been removed promptly and antibiotic therapy given.